The Management of Acute Abdominal Pain in Children with Sickle Cell Disease

Document Detail	
Document Type	Clinical Guideline
Document name	STSN Guidelines for the management of acute abdominal pain in children with sickle cell disease
Version	V1
Effective from	22 February 2022
Review date	22 February 2024
Authors	STSTN paediatrics clinical guidelines group
Superseded documents	
Keywords	Sickle cell disease,

Change History		
Date	Change details, since approved	Approved by

Table of Contents

Introduction

Acute abdominal pain is a common reason for admission to hospital in children with sickle cell disease (SCD). It may be related directly to vaso-occlusion, be a complication of treatment or unrelated to SCD. This guideline is aimed at all children with sickle cell disease (SCD) and acute abdominal pain. It covers the commonest causes of abdominal pain, suggests further investigations and when paediatric surgical involvement should be sought

Target Audience

Paediatricians, Paediatric Haematologists, Paediatric Surgeons and Paediatric Nursing staff treating children with sickle cell disease

Key changes from previous guideline

Yes

Same title as last version

- Addition of deferasirox as possible cause of abdominal pain
- Removal of antibiotic doses
- Addition of Testicular torsion as cause of pain and in exam section
- PIMS-TS as cause of abdominal pain – in the differential diagnosis of abdominal pain but to be aware of the possible pitfalls in SCD due to raised inflammatory markers in acute episodes – caution in making this diagnosis – discussion with senior colleagues in haematology, paediatrics and surgery.
- Incentive spirometry
- VTE prophylaxis for postpubertal patients immobile in bed

Derogation from NICE Guidance or Royal Marsden Manual

NA

Indications & Contra-indications

Applicable to all children with sickle cell disease who present with abdominal pain.

Treatment & Management

The different types of interventions and treatments to be included and excluded – for example, diagnostic tests, surgical treatments, medical and psychological therapies, rehabilitation and lifestyle advice.

The patient management algorithm of a particular disease/condition.

The Management of Acute Abdominal Pain in Children with Sickle Cell Disease

Background

Acute abdominal pain is a common reason for admission to hospital in children with sickle cell disease (SCD). It may be related directly to vaso-occlusion, be a complication of treatment or unrelated to SCD. This guideline is aimed at all children with sickle cell disease (SCD) and acute abdominal pain. The guideline applies to all patients with SCD who are currently under the care of the Paediatric team. It is mainly aimed at being a tool for the medical team managing these patients, but any member of the multidisciplinary team may find it useful.

Contents of guideline

Aim

To provide guidance on the management of children with sickle cell disease admitted with abdominal pain. It is mainly aimed at being a tool for the medical team managing these patients, but any member of the multidisciplinary team may find it useful.

Causes of Acute Abdominal Pain

Acute abdominal pain is a common symptom in children with sickle cell disease. Possible causes include:

Common

- Constipation with faecal impaction
- Acute abdominal sickling (also called abdominal painful crisis, girdle syndrome)
- Gastroenteritis
- Urinary tract infection

Less Common

- Cholecystitis
- Pancreatitis
- Parvovirus B19 infection
- Splenic sequestration
- Hepatic sequestration
- Acute appendicitis
- Acute chest syndrome
- Gastritis/peptic ulcer disease
- Septicaemia

Rare

- Hepatic infarction
- Viral hepatitis
- Ischaemic cholangiopathy
- Ischaemic colitis
- Renal papillary necrosis
- Testicular torsion in boys
- COVID-19 PIMS-TS

Urgent Management and Assessment

If the child is in pain, analgesia should be given according to the Pain Guidelines (INSERT LINK to local guideline if applicable)). If clinical assessment and Early Warning Score indicate the child is shocked or collapsed they should be urgently resuscitated according to standard protocols.
History should initially focus on identifying the more common diagnoses including:

- Does pain feel like previous episodes of pain due to SCD?
- Has the child been constipated or recently using codeine (or dihydrocodeine if <12 years)?
- Has the child been taking non-steroidal anti-inflammatory drugs frequently?
- Is there diarrhoea or vomiting?
- Are there features suggestive of dehydration/reduced urine output
- Is there a history of gall stones or cholecystitis?
- Has jaundice increased?
- Are there symptoms of a urinary tract infection?
- Is there a history of splenomegaly or splenic sequestration?
- Are they taking iron chelation (Exjade FCT – Deferasirox)?
- History of COVID-19 infection? (see below)

Examination

Full examination should be performed, with particular reference to:

- Bowel sounds: in severe abdominal vaso-occlusion, bowel sounds are usually absent, but a silent abdomen also raises the possibility of acute surgical complications, including perforation.
- Signs of appendicitis
- Signs of cholecystitis
- Splenomegaly
- Hepatic enlargement or tenderness
- Testicular examination in boys
- Signs of shock, cardiac failure

Initial Investigations – please compare to steady state values

- FBC, U&E's, reticulocyte count
- Blood Group and antibody screen
- CRP
- Blood gas and Lactate
- Liver function tests including ALT and LDH
- Serum amylase and lipase
- Blood cultures if temperature >38^{0}C
- Urine for microscopy and culture
- Pulse oximetry on air
- Stool culture if diarrhoea
- Malaria screen if travel to an endemic area

- COVID-19 PCR or COVID-19 antibodies if history of previous infection/exposure.
- if considering Paediatric Inflammatory Multi-system Syndrome (PIMS-TS) in the differential diagnosis of abdominal pain, exercise caution since many of the diagnostic criteria for PIMS-TS are based on raised inflammatory markers which are commonly found in severe acute sickle vaso-occlusive episodes eg ferritin, CRP, fibrinogen. Please discuss these cases with senior medical staff (paediatric/paediatric haematology/paediatric surgery).

Imaging – consider -

- Ultrasound of the liver, gall bladder, pancreas or spleen if there is evidence of cholecystitis, pancreatitis, hepatic enlargement or tenderness, splenic enlargement or tenderness.
- Chest X-ray if there is chest pain, hypoxia, chest signs, or an acute abdomen and to assess for the presence of air under the diaphragm.
- Abdominal X-ray if acute abdomen (following discussion with paediatric surgeons). This may also show evidence of constipation.
- Ultrasound of kidneys, ureters and bladder if there is haematuria, renal colic or severe lower abdominal pain.

Initial Management

The child's condition should be stabilised and appropriate analgesia given.

- Intravenous fluids should be started if the pain is severe, there are no bowel sounds or if there is diarrhoea or vomiting. Normal full maintenance fluids should be started; however, if the child is assessed as being clinically dehydrated, the rate should be increased guided by frequent clinical assessment, with careful monitoring and fluid balance. It is important to avoid fluid overload, especially if there are associated respiratory symptoms.
- Please see the fluid guideline for guidance on volume of fluids of normal maintenance and monitor fluid balance and renal function.
- If there are no bowel sounds or a surgical cause such as acute appendicitis is suspected, the child should be made nil-by-mouth and the paediatric surgery team asked to see the patient urgently. Consider NGT placement if clinically indicated.
- Penicillin V should be continued at the prophylactic dose unless the temperature is >38^{0}C or there are no bowel sounds; in which case intravenous antibiotics should be started as per local protocol, to provide cover for gram negative organisms, taking into account potential penicillin allergy and /or G6PD deficiency (see table below).
- If the patient is taking hydroxyurea, it should be continued unless the patient is nil-by-mouth, the blood tests show evidence of toxicity (neutrophils < 1.0 x10^9/l, platelets < 80 x10^9/l, reticulocytes <80 x10^9/l, ≥ 50% increase in serum creatinine, ALT > 110 IU/l).
- Incentive spirometry should be started according to ward protocols.

Empirical antibiotics for acute abdominal pain and temperature > 38^{0}C in a child with sickle cell disease
Refer to BNFc for dosing
Traffic light system for Penicillin Allergy:
Drugs in RED are contraindicated in Penicillin Allergy

Drugs in YELLOW should be prescribed with caution and only if penicillin allergy is lsess severe – administer under close supervision **Drugs in GREEN** are considered safe.	
1st line empirical antibiotic **Temp >38°C** **Absent bowel sounds** **And if neutropenic**	**Piperacillin-tazobactam (Tazocin) IV TDS** **Piperacillin-tazobactam (Tazocin) IV TDS** **PLUS** **Gentamicin IV OD** (adjusted according to plasma concentration)
If either - History of ESBL-producing Gram negatives - History of recent broad spectrum antibiotic use	**Meropenem IV TDS** **review within 72 hours with culture results**
Penicillin anaphylaxis	Vancomycin IV (adjusted according to plasma concentration) Ciprofloxacin IV (avoid in G6PD deficiency) Metronidazole IV
Known MRSA colonisation	Not relevant in this clinical presentation
Treatment duration	7-10 days with clinical review If cultures remain negative consider stopping after 5 days

Further Investigations

Further investigations may be appropriate depending on the initial findings:

- Parvovirus B19 IgM and IgG serology if the reticulocyte count is < 100 x10^9/l.
- Hepatitis serology if the ALT is >110 IU/l
- CT abdomen may be indicated after surgical review

Further Specific Management of Common Causes

This will depend on the initial diagnosis and the surgical team may advise additional imaging.

Acute abdominal vaso-occlusive episode

- Children with a vaso-occlusive episode involving the abdomen are often in severe pain and may require PCA or NCA analgesia according to local protocols, with close monitoring of pain score and sedation.
- The patient should be re-examined frequently to ensure that the pain is being controlled and that an underlying cause of the pain is not becoming apparent, i.e. acute surgical conditions.

Re-examination should include chest examination and assessment of oxygen requirement, to ensure that acute chest syndrome is not developing; this is more likely when patients are lying immobile in bed, and when pain causes abdominal splinting and reduced respiratory movement. Examination should be repeated in the first few hours and 6-12 hourly thereafter depending on progress.

- Early discussion with tertiary centre paediatric surgical and haematology teams should be considered for deteriorating patients and those at high likelihood of requiring surgery.
- In general conservative management is followed unless there is a definite surgical diagnosis such as acute appendicitis or perforation. If a patient requires urgent abdominal surgery eg for appendicitis/perforation, they must be discussed with the consultant paediatrician/haematologist, consultant anaesthetist and consultant surgeon. The haematology team will advise about perioperative management including the need for blood transfusion, especially if the Hb <90g/l.. If the patient has a history of severe sickle cell complications, eg acute chest syndrome requiring PICU support, they should be discussed with HDU/PICU to plan a postoperative bed.
- Blood tests should be performed daily to monitor renal and hepatic function and FBC. If the haemoglobin falls >20g/l below the steady-state, or below 50g/l, top-up blood transfusion will usually be necessary.

Constipation

- Typically the pain is moderate or mild, with an intermittent or colicky nature. Usually there is a history of infrequent or painful defaecation.
- Appropriate analgesia should be given, depending on the severity of the pain. Paracetamol and ibuprofen are typically sufficient, although opiates are sometimes needed.
- The child should be encouraged to drink.
- Laxative therapy such as macrogol should be started, and lactulose and senna added as necessary.
- An enema may be appropriate.

Cholecystitis

- This is usually diagnosed with a combination of typical symptoms and signs, increased bilirubin, and gallstones with a thickened gall bladder on ultrasound examination.
- The surgical team should be involved in the care of the patient at an early stage and the paediatric hepatology team at Kings College Hospital should be informed if acute cholecystitis is diagnosed.
- If vomiting is severe or bilious a nasogastric tube may be necessary in addition to nil-by-mouth, intravenous fluids and intravenous antibiotics (to ensure adequate gram negative cover).
- Management of the acute episode is typically conservative.
- After one episode of cholecystitis, an elective cholecystectomy will typically be performed after recovery from the acute episode, depending on surgical considerations and the views of the patient/family, since it is likely to recur if gallstones are present.
- Further investigations such as ERCP will be organised by the paediatric hepatology team.

- At discharge it should be ensured that the patient is referred for review with the surgical team.

Other diagnoses

- Other diagnoses should be managed according to standard protocols.
- Acute vaso-occlusive pain and acute chest syndrome can develop during and complicate any admission for unrelated reasons, such as gastroenteritis, or urinary tract infection.
- In general, children with sickle cell disease and abdominal pain should be monitored closely, with particular attention to fluid balance, early treatment of possible infection, and early investigation of any respiratory or neurological symptoms or signs.
- If PIMS-TS is considered in the differential diagnosis please discuss with senior medical staff – there is considerable overlap some of the diagnostic tests identifying increased inflammation which are also seen in acute severe vaso-occlusive episodes.

Supportive care

- Accurate fluid balance
- Incentive spirometry
- Analgesia – PCA/NCA may be necessary
- Maintenance of oxygen saturations >97% - development of an oxygen requirement should prompt urgent review and CXR for possible acute chest syndrome
- Postpubertal patients – consider LMWT heparin VTE prophylaxis if immobile in bed

Reference

Ahmed S, Shahid RK & Russo LA. Unusual causes of abdominal pain: sickle cell anaemia. *Best Practice and Research Clinical Gastroenterology* 2005; 19, 297-310.

Contact details:

KCH Paed sickle CNS: 020 3299 4752
KCH Paed sickle consultants: david.rees2@nhs.net; subarna.chakravorty@nhs.net sue.height@nhs.net; john.brewin@nhs.net
KCH paediatric haematology SPR: mobile extension 39283
GSTT Paed sickle CNS: 020 7188 9432
GST Paed sickle consultants: samah.babiker@gstt,nhs.uk; Nick.Fordham@gstt.nhs.uk
GSTT paediatric haematology SPR: Bleep 1621
QEW Paed sickle CNS: 07741233556
QEW Paed sickle consultants: julie.lord@nhs.net; aruj.qayum@nhs.net
UHL Paed sickle CNS: 07741233556
UHL Paed sickle consultants: s.wilkinson6@nhs.net
CUH Paeds sickle CNS: 0208 2517229
CUH Paeds sickle consultants: nazmachowdhury@nhs.net

Additional contacts can be found on the STSTN website (www.ststn.co.uk)

Guidelines written by the STSTN paediatric writing group:

Dr Sue Height
Dr Sarah Wilkinson
Dr Rachel Kesse-Adu
Professor Baba Inusa
Dr Samah Babiker
Dr John Brewing
Dr Helen Oram